Sarcoidosis of the Spinal Cord: Literature Review and Report of Eight Cases

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Sarcoidosis, which affects African Americans more than it does other racial/ethnic groups, only rarely manifests initially as spinal cord dysfunction. This paper presents the findings of eight patients with spinal cord dysfunction as part of a presentation of sarcoidosis. After reviewing these cases, we devised an algorithm to diagnose and manage spinal cord sarcoidosis.

Key words: spinal cord ■ sarcoidosis ■ myelopathy ■ cauda equina ■ neurosarcoidosis

INTRODUCTION

Sarcoidosis of the spinal cord is rare with an incidence estimated at 0.43%.[1] The information available about the spinal cord sarcoidosis comes from scattered case reports, small series and expert opinions. We reviewed the English literature over the past 65 years and added eight cases that reflect the difficulties in diagnosing and managing this unusual disease.

METHODS

A Medline search of the English literature published between 1954 and December 2004 was performed using the following terms: "spinal cord sarcoidosis," "spinal sarcoidosis," "myelopathy" and "sarcoidosis," "cauda equina" and "sarcoidosis," and "neurosarcoidosis." Articles were also included from the references section of the reviewed literature up to to 1940. Cases were included if at least the age was reported in the case description. We here present the following eight patients (Table 1). They represent our experience in a tertiary referral sarcoidosis clinic over the past 10 years.

Case 1

In December 2002, a 44-year-old man developed tingling in both arms and feet. Gadolinium-enhanced MRI of the brain and spinal cord showed enhancement of the leptomeninges of the posterior fossa, basal cisterns of the supratentorial compartment, upper cervical spine and the cauda equina. Biopsy of an intradural extramedullary C7 mass showed nonnecrotizing granulomas (Figure 1). Stains and cultures were negative for mycobacteria, fungi and parasites. Cerebrospinal fluid (CSF) analysis was normal. No other organ was affected. Serum angiotensin-converting enzyme (ACE) level was normal. Prednisone 80 mg/day induced rapid improvement; after four months, prednisone was reduced to 5 mg/day. Steroid-induced side effects mandated a change in drug regimen to hydroxychloroquine 200 mg twice daily, reduced to 200 mg daily in December 2003. Repeat MRI of the brain and spine showed marked decrease in enhancement

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of the lesions. The patient remains in clinical remission, having been followed to December 2003.

Case 2

In June 1996, a 47-year-old man complained of fever, dyspnea, skin rash and weight loss. An extensive work-up included biopsies of the liver and stomach, both consistent with sarcoidosis. Shortly after, he developed leg weakness and dysesthesias, sexual dysfunction, and bladder and bowel problems. A burning sensation at the lower part of his torso plagued him. MRI of the spine showed increased attenuation at T4–T5. Prednisone 60 mg/day, improved the symptoms and was later reduced to 5 mg/day. After neurological symptoms recurred, prednisone was increased to 40 mg/day. By April 1998, symptomatic control was achieved, and prednisone was reduced slowly to 5 mg/day; hydroxychloroquine 200 mg twice a day was added. Repeat MRI in November 1998 was normal, and the patient was asymptomatic; all drug therapy was tapered off and discontinued. In March 2001, after elevated creatine kinase levels were noted, a muscle biopsy showed granulomas consistent with sarcoidosis; prednisone and hydroxychloroquine were restarted.

Case 3

In April 2004, a 52-year-old woman developed progressive lower extremity weakness. MRI showed an abnormal signal extending from C6–T7 without abnormal enhancement and high signal intensity in a periventricular distribution. CSF showed 164 mononuclear cells and protein of 660 mg/dl. There were no oligoclonal bands; viral cultures were negative. A provisional diagnosis of transverse myelitis was made, and she was given methylprednisolone for three days followed with a tapering course of oral steroids and rehabilitation over two weeks. In August 2004, she was readmitted to the hospital with worsening leg weakness, ataxia, urinary retention and episodes of expressive dysphasia. MRI with gadolinium showed extensive arachnoid enhancement involving the convexities, sylvian fissures, basal cisterns and cerebellar folia. Confluent periventricular high signal change was noted. The spinal cord showed stable high signal areas with the main ones being at T3–T4 level. CT of the chest and abdomen showed mediastinal lymphadenopathy; thoracoscopic lymph node biopsy showed sarcoidal granulomas. Stains and cultures for fungi and acid-fast bacilli were negative. Prednisone 80 mg/day was started, and she was sent to rehabilitation.

Case 4

In November 2003, a 53-year-old man developed bilateral ankle swelling. A chest radiograph showed hilar lymphadenopathy. He was given prednisone. In January 2004, he started complaining of lower extremity weakness, paresthesia, inability to ejaculate and frequent urination. MRI showed edema of the spinal cord at T1–T8 and enhancement of the leptomeninges at T5–T6. Computed tomography (CT) of the chest showed hilar lymphadenopathy. CSF examination showed lymphocytic pleocytosis, elevated protein, normal glucose and normal cytology. A cervical lymph node biopsy showed noncaseating granuloma. Prednisone, hydroxychloroquine and methotrexate were administered. In November 2004, a follow-up MRI of the spine showed no reactivation of the disease.

Case 5

In July 2001, a 41-year-old Caucasian man noted numbness and weakness of the left leg, right arm and the right leg. In January 2002, an MRI with gadolinium contrast showed an intramedullary cord mass involving C5–T1 with enhancement. He was started on methylprednisolone with transient improvement; however, the diagnosis was never established. In March 2002, his symptoms worsened, and a repeat MRI showed worsening of the cord expansion and extension into the upper thoracic spine. In April 2002, he had a C3–C7 laminectomy for decompression and resection of the cervical mass; biopsy showed noncaseating granulomas (Figure 2). He was given prednisone 80 mg/day with improvement in his symptoms. He gained weight. Methotrexate was added in an attempt to taper the prednisone. In June 2003, the patient developed a band-like sensation around his waist. MRI revealed an enhancing 1.5-cm mass at the level of C7. He was given intravenous methylprednisolone at 1,000 mg/day for three days followed by prednisone at 80 mg/day with improvement in muscle power. An MRI examination showed a decrease in the size of the lesion; shortly after, prednisone was reduced to 50 mg/day, and his symptoms started to worsen. Methotrexate 15 mg/week was started. In January 2004, he developed sudden low back pain, paraplegia and urinary retention; MRI showed a T12 vertebral body fracture compressing the cord.

Case 6

In June1998, a 70-year-old Caucasian man suffered from tunnel vision and swelling of the optic nerve. CSF analysis showed lymphocytosis and elevated proteins. A brain MRI showed abnormal enhancement of the pia of the brainstem and of several cranial nerves, including the optic nerves. This subsided with supportive therapy. A few months later, he started to suffer from a tingling sensation in the arms, shoulder and neck, and quadriparesis. An MRI of the spinal cord revealed an intramedullary enhancing mass at C1 (Figure 3). He was given dexamethasone with improvement in his symptoms. A search for other organ involvement revealed a right superior medi-

Table 1. Features of our reported cases

Patient/Age/ Sex/Race	Level of Spinal Cord on MRI	Other Organ Involvement	Spinal Cord Biopsy	Other Organ Biopsy	Laboratory Findings	Treatment	Outcome	Duration of Follow-Up
1/44/M/AA	C7	Brain	Yes	No	CSF: NL, ACE: NL	Prednisone then added hydroxy-chloroquine	Improvement clinical and on MRI	1 year
2/47/M/AA	T4–T5	Skin, liver, muscle	No	Liver, muscle	NA	Prednisone then added hydroxy-chloroquine	Improvement then relapse when stopped prednisone. Improvement when prednisone restarted	5 1/2 years
3/52/F/NA	C7–T7	Brain, mediastinal lymph nodes	No	Mediastinal lymph nodes	CSF: pleocytosis, elevated protein	Prednisone	Improvement	4 months
4/53/M/AA	T1–T8	Mediastinal and cervical lymph nodes	No	Cervical lymph nodes	CSF: pleocytosis, elevated protein	Prednisone then added hydroxy-chloroquine and methotrexate	Improvement	2 years
5/41/M/W	C5–T1	No	Yes	No	NA	Prednisone, laminectomy, methotrexate	Complications from surgery followed by remissions and relapses	2 1/2 years
6/70/M/W	C1	Brain, mediastinal lymph nodes	No	Mediastinal lymph nodes (necrotizing sarcoidosis)	CSF: pleocytosis, elevated proteins	Prednisone, hydroxy-chloroquine, anti-TB meds	Improvement	3 1/2 years
7/31/M/NA	T4	Hilar lymph nodes	No	Hilar	ACE: elevated	Prednisone	Improvement	2 years
8/41/M/NA	T1–T12	Hilar lymph nodes	No	Lungs	NA	Prednisone, hydroxy-chloroquine, methotrexate	Improvement	7 years

M: male, F: female, AA: African-American, NA: not available, W: white, c: cervical, t: thoracic, CSF: cerebrospinal fluid, ACE: angiotensin-converting enzyme, TB: tuberculosis

astinal mass that was removed in May 1998. Biopsy of the mass showed necrotizing and nonnecrotizing granulomas. No acid-fast or fungal organisms were observed either by direct examination or culture. In view of the mixed granulomatous response and a positive tuberculin test, he was given antituberculous treatment. Follow-up MRIs in October1999 and January 2000 continued to show enhancement in the cervical cord. The diagnosis of the spinal cord sarcoidosis was entertained and he was given prednisone 80 mg/day. He improved significantly, and prednisone was tapered. Hydroxychloroquine 200 mg twice daily was started. The antituberculous medications were discontinued. In April 2000, a repeat MRI showed marked improvement. In October 2000, he was taken off prednisone. When last seen in January 2002, he was stable on hydroxychloroquine 200 mg/day.

Case 7

In September 1994, a 33-year-old man suffered from flu-like symptoms. A chest radiograph showed bilateral hilar and mediastinal lymphadenopathy. A biopsy of the node showed noncaseating granulomas. No treatment was given. In April 1996, he developed severe leg weakness, ataxia and exaggerated motor reflexes. MRI of the spine revealed an enhancing 1-cm lesion in the spinal cord at T4 level. His serum ACE level was elevated at 228 mg/l. He was started on prednisone 80 mg/day, with symptomatic improvement within two weeks. After four weeks, a repeat MRI of the spine showed improvement. He continued to improve, and his prednisone was tapered down by 10 mg every week to a maintenance dose of 10 mg/day. When last seen in September 1996, he was asymptomatic and not on any treatment.

Case 8

In September 1993, a 41-year-old man presented with dyspnea and fatigue. Chest x-ray showed bilateral hilar adenopathy, and a transbronchial biopsy showed noncaseating granulomas. He was given prednisone. He improved, and prednisone was subsequently tapered off. In December 1996, he complained of numbness of the lower half of his body and lower extremity weakness. MRI showed multiple enhancing masses of the entire thoracic spine (Figure 4). The diagnosis of probable spinal sarcoidosis was made, and he was started on prednisone with clinical and radiological improvement. In June 1999, he returned with recurrent lower extremity weakness with worsening of his MRI findings. He was restarted on prednisone and hydroxychloroquine. He improved minimally. Hydroxychloroquine was stopped and methotrexate started. On this regimen, he improved, and a follow-up MRI showed decrease in the size and enhancement of the lesions.

RESULTS

One-hundred-sixty-four cases of spinal cord sarcoidosis were identified from case reports and case series in the literature.[1-115] We added eight cases of our own to bring the total to 172.

Patient Characteristics

The characteristics and clinical presentation of all the patients reviewed are summarized (Table 2). Ninety-five (55.6%) of 172 patients were men. The group comprised 52 (61.9%) African Americans, 26 (30.9%) Caucasians, four (3.6%) Asians and three (3.6%) members of other ethnicity. The age distribution ranged from 17–71 years, with a mean of 42.8 years.

Clinical Presentation

In 56.9%, spinal cord disease was the presenting manifestation of sarcoidosis, and in 15.6% of the patients it was the only manifestation of disease. The cervical spine was affected in 93 (61.4%), the thoracic spine in 87 (58.1%), the lumbar spine in 19 (12.1%) and the cauda equina in 19 (12.8%) patients. In 49 (32.7%) patients, more than one area of the cord was affected (Table 2). The lungs were the most commonly affected extra-axial organ (58.2%) in patients whose

Figure 1. Noncaseating granuloma in an intramedullary spinal cord mass (case 1) (H&E x100)

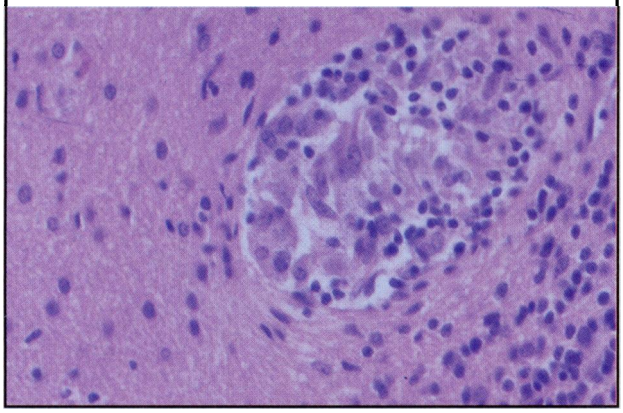
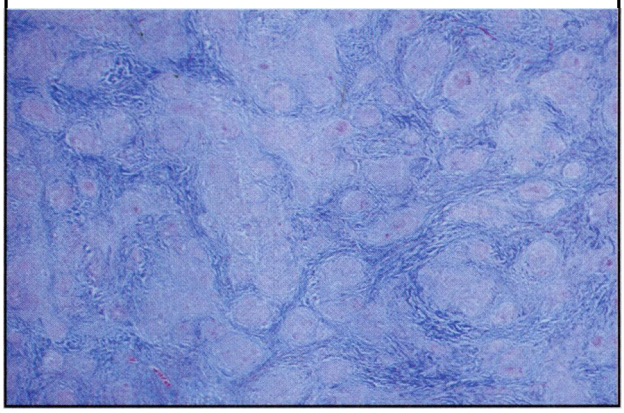


Figure 2. Spinal cord granuloma in case 5. (Trichrome stain x40)



first manifestation of sarcoidosis was the spinal cord disease. Table 3 shows the proportion of organ involvement in these patients.

Diagnosis

The following criteria were used to define the diagnosis of sarcoidosis of the spinal cord.[31]

1. **Definite:** clinical presentation compatible with neurosarcoidosis (of the spine), exclusion of other possible causes, and positive nervous system histology (biopsy or autopsy). Ninety-one (52.9%) of 172 cases were considered to have definite diagnosis of spinal sarcoidosis.
2. **Probable:** clinical presentation compatible with neurosarcoidosis (of the spine), laboratory support of CNS inflammation, exclusion of other possible causes and evidence of systemic sarcoidosis. Sixty (34.9%) cases were classified as probable sarcoidosis.
3. **Possible:** clinical presentation compatible with neurosarcoidosis (of the spine) and exclusion of other possible causes. Twenty-one (12.2%) met possible criteria.

Table 2. Patient characteristics and clinical presentations

Characteristic	Number
Age	Mean 42.8 years (range 17–71)
Sex	
Male	55.2% (n=95)
Female	44.8% (n=77)
Ethnicity	
African American	61.9% (n=52)
Caucasian	30.9% (n=26)
Asian	3.6% (n=3)
Other	3.6% (n=3)
Spinal cord affection	
First manifestation of disease	56.9% (n=98)
Only manifestation of disease	15.6% (n=27)
Location of spinal disease	
Cervical	61.4% (n=91)
Thoracic	58.1% (n=86)
Lumbar	12.1% (n=18)
Cauda equina	12.8% (n=19)
Multiple levels	32.4% (n=48)
Diagnosis	
Definite	
Spinal cord biopsy	39.5% (n=68)
Autopsy	13.4% (n=23)
Total	52.9% (n=91)
Probable	34.9% (n=60)
Possible	12.2% (n=21)
ACE level (serum) (n=72)	
Elevated	34.7% (n=25)
Normal	65.3% (n=47)
CSF Analysis (n=122)	
High protein	80.3% (n=98)
Pleocytosis	69.7% (n=85)
Low glucose	12.3% (n=15)

ACE: angiotensin-converting enzyme, CSF: cerebrospinal fluid

Table 3. Organ involvement in patients with spinal cord disease as the first manifestation of sarcoidosis

Organ Involvement	Number of Patients (Total=98)	Percentage
None	27	27.6%
Lung	57	58.2%
Eye	10	10.2%
Liver	6	6.1%
Lymphadenopathy (extrathoracic)	5	5.1%
Heart	4	4.1%
Skin	4	4.1%

Serum ACE level was reported in 72 (41.9%) of cases. It was elevated in 25 (34.7%) of reported cases and normal in 47 (65.3%).

CSF analysis was performed in 122 (70.1%) of cases. The most common abnormality was elevated protein level in 98 (80.3%) followed by pleocytosis 85 (69.7%) and low glucose in 15 (12.3%). Elevated IgG level was reported in 6.6% of cases.

Magnetic resonance imaging was the most common test used to diagnose spinal cord affection. Swelling of the cord and enhancement with gadolinium were the characteristic, although nonspecific, findings.

Treatment

Steroids were the drug most commonly used in 129 (75%) of 172 cases. Fifty-five (32%) of the patients received only prednisone, whereas the rest received various combinations. Other drugs used included methotrexate in six (3.5%), chloroquine in one (0.6%), hydroxychloroquine in eight (4.7%), cyclophosphamide in three (1.7%), and infliximab in two patients. One patient received radiotherapy.

Laminectomy, in conjunction with corticosteroids, was used in 65 patients (37.7%), whereas in nine (5.2%) cases, patients received laminectomy without any additional medical treatment.

Outcome

Information about outcome was available on 138 patients. Ninety-one (65.9%) patients improved, 34 (24.6%) deteriorated and 13 (9.5%) remained stable.

DISCUSSION

Sarcoidosis is a multisystem granulomatous disease of unknown etiology that most commonly affects the lungs, eyes and skin. Because spinal cord sarcoidosis is rare, little is known about its diagnosis and management.

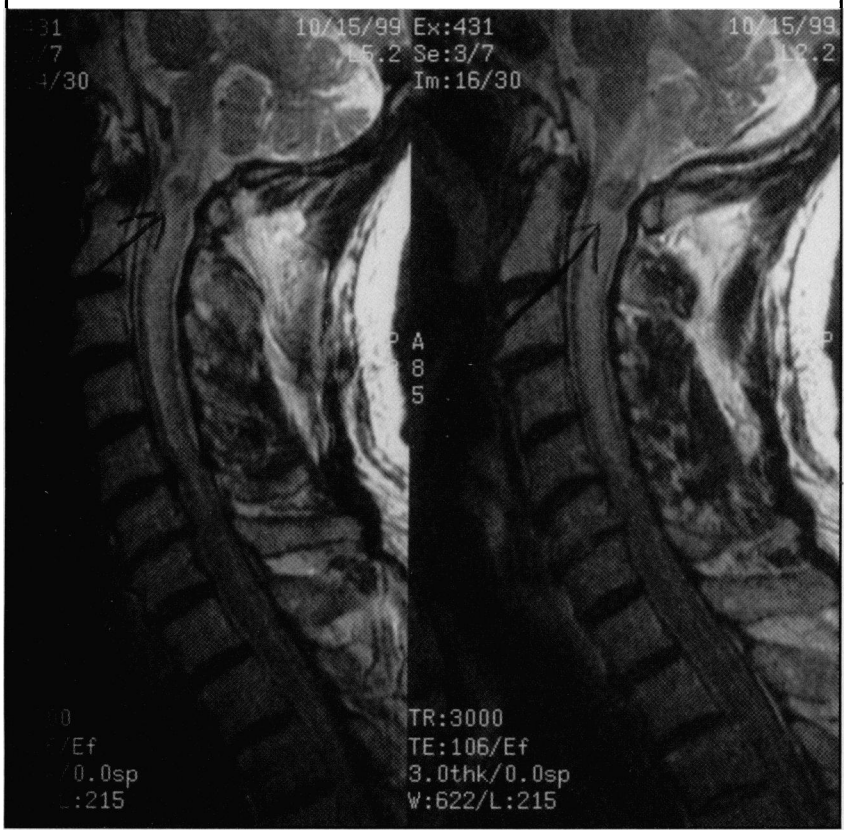
Epidemiology

Sarcoidosis occurs worldwide. It affects both sexes, but most studies suggest a slightly higher disease rate for women.[116] It has a predilection for adults aged <40 years, and in Scandinavia and Japan a second peak occurs in women age >50.[117] In the United States, the lifetime risk of sarcoidosis is 0.85% for Caucasians and 2.4% for African Americans, who are more severely affected.[118]

Five-to-17% of patients with multisystem disease will have clinical evidence of CNS involvement.[72,94] Autopsy studies, however, estimate that 50% of patients have subclinical involvement of the nervous system.[119] Spinal cord sarcoidosis occurs in about 0.43% of patients.[1] It affects more men than women and more African Americans than other ethnic/ racial groups. The peak incidence is 42.9 years.

Spinal cord involvement can occur as a part of multisystem sarcoidosis either as the first manifestation or later in the course of the disease. The spinal cord may be the only organ affected with the disease, as happened in 27 (15.6%) of the patients reported in the literature and in two of our eight patients. Spinal cord sarcoidosis as the first manifestation of multisystem disease is most often followed first by lung involvement, then by ocular symptomatology.

Figure 3. Magnetic resonance image of a localized intramedullary lesion at level of C7 (case 6)



Diagnosis

Clinical presentation. Spinal cord sarcoidosis may present with paraparesis,[67] quadriparesis,[75] sensory changes, autonomic dysreflexia,[115] radicular symptoms or cauda equina syndrome.[6,81] The cervical cord is affected more than other segments of the spinal cord.[74] In a few patients, the cord is diffusely involved, encompassing all the segments (32.6%).

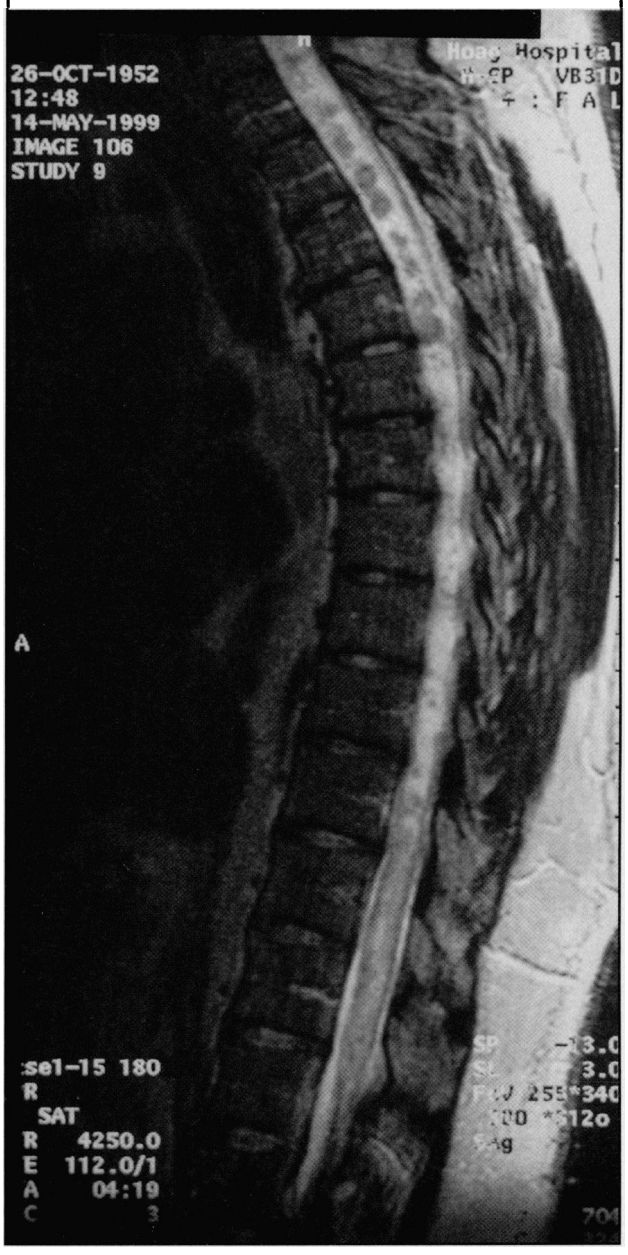
The clinical conditions considered in the differential diagnosis are spinal cord tumors, such as astrocytoma and ependymoma, multiple sclerosis, Lyme disease, postinfectious encephalomyelitis, tuberculosis, lymphoma, metastatic tumors, paraneoplastic processes[32], Guillain-Barre syndrome[73,102] and vasculitides.[120]

Neuroimaging. MRI is very sensitive to localize the lesions; lesions enhance with gadolinium except late in the disease, when cord atrophy occurs.[53] Appearances are not specific and highly variable.[121] Enhancement was seen to start from the meninges towards the cord parenchyma, and it led to the speculation that sarcoidosis starts at the meninges and then affects the cord.[64] Junger et al.[53] classified the MRI manifestations of the intramedullary lesions into four stages. They postulate the presence of a correlation between gadolinium enhancement and inflammation. Stage 1 presents as linear leptomeningeal enhancement and may present early inflammation. In stage 2, the inflammation spreads through the perivascular (Virchow-Robin) spaces leading to parenchymal involvement and presents as diffusely enhancing lesions with cord enlargement. Stage 3 presents with relatively normal-size cord with focal or multifocal enhancement. In stage 4, there is relative atrophy of the cord and no enhancement. MRI is also helpful in detecting response to treatment[122,123] and is a useful marker to detect subclinical recurrence.[11,24]

Cerebrospinal fluid analysis. Pleocytosis (especially lymphocytic), elevated proteins and sometimes low glucose are the most common abnormalities. However, this may be seen in other disorders such as multiple sclerosis and systemic lupus erythematosus.[120] The CSF ACE levels may be elevated in 50% of patients with neurosarcoidosis. This may also be elevated in other central CNS infections and malignant tumors.[124] The ACE level in CSF changes with disease activity[125,126] and may be useful in disease monitoring. The level of ACE in the CSF does not correlate with that of the serum.[124] CSF lysozyme and beta-2 microglobulin levels are elevated in 75% and 68% of neurosarcoidosis patients. This was also observed in patients with meningitis and CNS tumors and, thus, they are not specific for sarcoidosis. Their levels in the CSF do not correlate with the serum levels.[126]

Role of biopsy. The diagnosis is suspected in patients with active systemic sarcoidosis; however, it is extremely difficult to make the diagnosis when spinal cord involvement is the first or sole manifestation of the disease. Biopsy of the spinal cord is an invasive procedure and is not without risk. The decision to obtain a biopsy of the spinal cord should be taken only after an extensive search for other organ involvement has been exhausted. This includes a thorough physical exam, radiography of the chest, laboratory work-up and possibly a gallium scan. Positron emission tomography (PET) has been used in a single report of neurosarcoidosis in search of other organ involvement.[12] In that report, the spinal cord lesions were hypermetabolic at the site of MRI abnormality, whereas the brain lesions were hypometabolic. The hypermetabolic hilar and mediastinal lymph nodes, which were biopsied, appeared normal on the chest radiograph but were enlarged on CT of the chest. Figure 5 shows a diagram for a suggested approach to diagnosis. In our review, the diagnosis was made by spinal cord biopsy or autopsy in 52.9% of the cases reviewed and by biopsy of other organs in 34.9%. Cases 1 and 5 illustrate the

Figure 4. T2-weighted magnetic resonance image showing multiple enhancing masses of the thoracic spine (case 8)

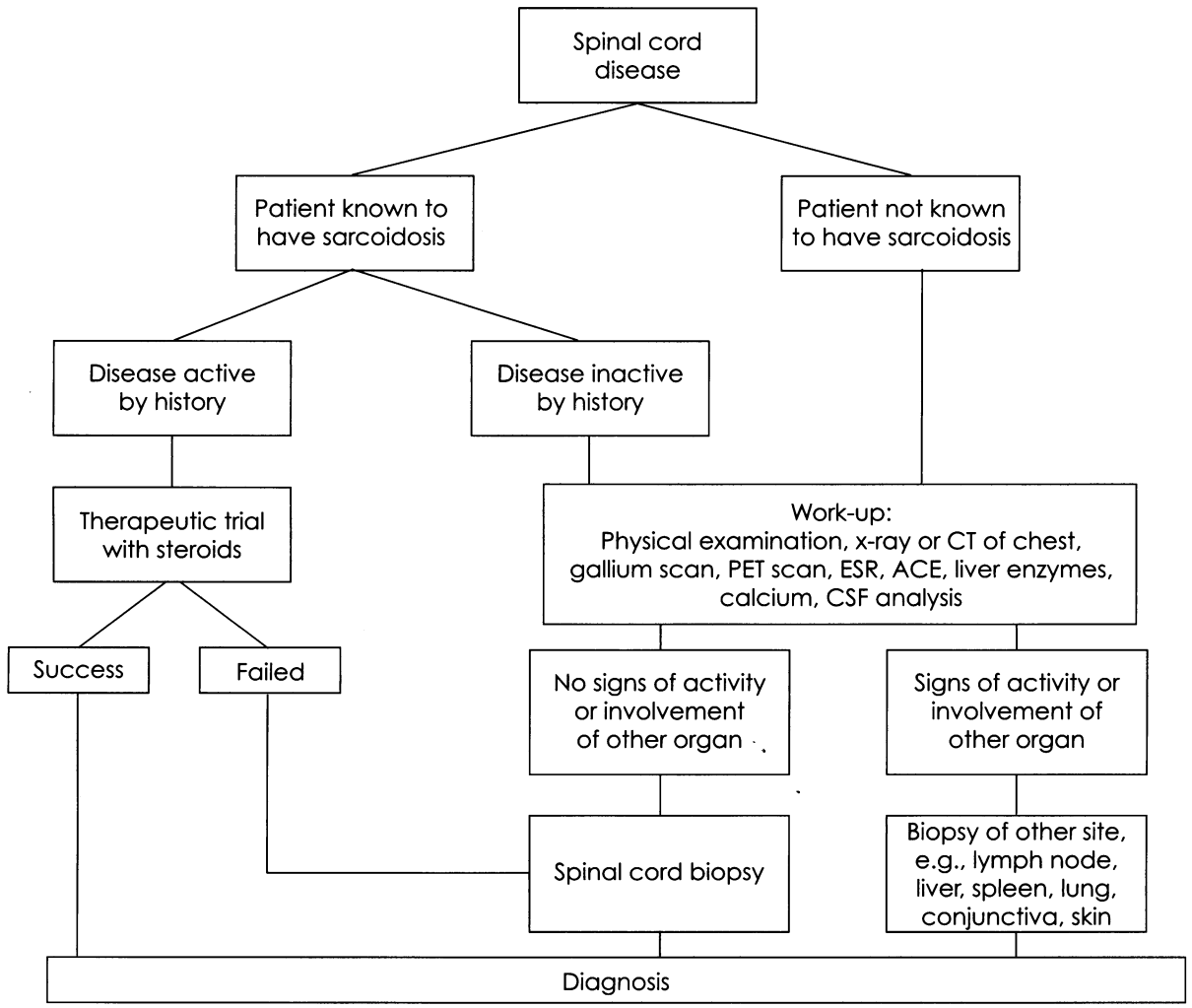


difficulty in making the diagnosis and thus the need to obtain a biopsy of the cord. Case 3 is an example of fruitful search of other organ involvement and avoidance to biopsy the spinal cord. In most of the cases, a biopsy was performed during a laminectomy procedure to relieve compression on the cord with or without an attempt to excise the mass. Attempting to excise the mass has been shown to result in poor outcomes,[111] as illustrated in case 5. It should be noted that misinterpretation of the frozen section biopsy of the cord has been reported and was speculated to occur because sarcoidosis is usually not a consideration before surgery, and spinal cord granulomas are smaller, less defined and associated with fewer giant cells than sarcoid lesions elsewhere.[64,96]

Treatment

Clear guidelines, based on double-blind prospective studies, for the treatment of spinal cord sarcoidosis do not exist. Corticosteroids are the agents of choice. In order to achieve rapid control of the disease, it may be necessary to give high-dose methylprednisolone intravenously for three days followed by prednisone at a dose of 1/mg/kg body weight daily.[120] Duration of therapy is often individualized. After 1–3 months, the patient is evaluated for response and side effects of therapy. Patients who do not respond by three months are unlikely to benefit from a more protracted course. The reasons for treatment failure include irreversible fibrotic disease, noncompliance, inadequate dosage and intrinsic corticosteroid resistance. Among steroid responders, the dose is slowly tapered to 5–10 mg/day or an every-other-day regi-

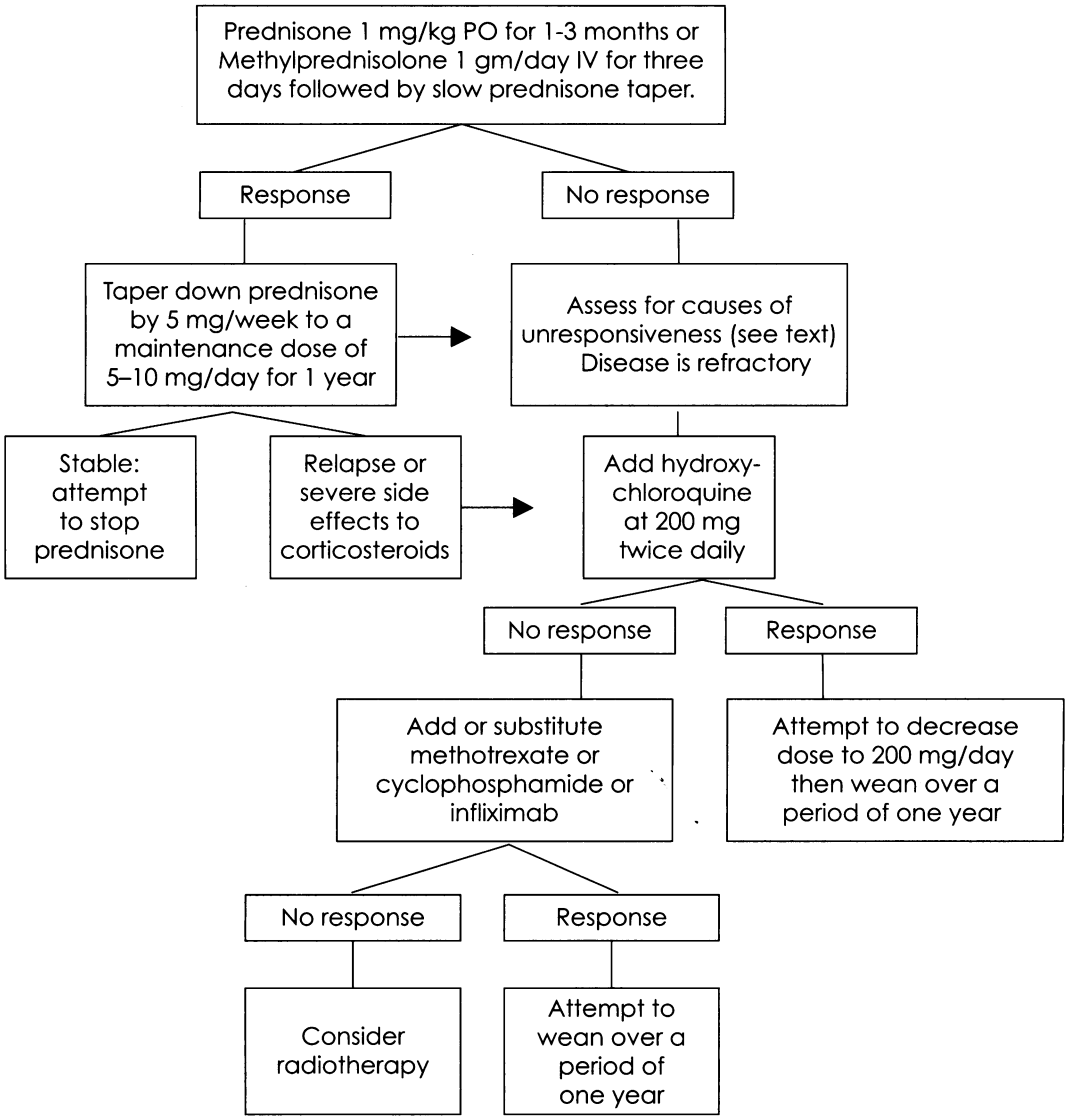
Figure 5. Diagnostic approach to patients with spinal cord sarcoidosis



CT: computed tomography, PET: Positron emission tomography, ESR: erythrocyte sedimentation syndrome, ACE: angiotensin-converting enzyme, CSF: cerebrospinal fluid

men. Treatment may need to be continued for a minimum of 12 months.[117] In general, after a favorable response achieving improvement, prednisone is tapered off to 5 mg/day. We recommend starting a second agent such as hydroxychloroquine while tapering down the steroid dose to maintain the patient in remission and to prevent the need to use a steroid for a long period with its resultant complications. Hydroxychloroquine at a dose of 200–400 mg/day and chloroquine 250 mg twice a day are effective in controlling refractory and chronic neurosarcoidosis.[127] Hydroxychloroquine is preferred to chloroquine; it is safer of the two.[117] It may be needed to add a cytotoxic agent because of severe side effects to corticosteroids. Methotrexate and azathioprine are the preferred agents for most patients.[117] Methotrexate is given orally or subcutaneously at 10–25 mg/week, and it was shown to have a response rate of 61% in neurosarcoidosis.[128] Routine use of folate supplementation at 1 mg/day is not recommended except for patients who develop side effects.[129] Azathioprine, given orally at 50 mg tid, has been used in chronic sarcoidosis with various results.[130,131] Cyclophosphamide is reserved for particularly difficult and refractory cases. Cyclophosphamide used in an intravenous weekly dose is effective in decreasing the dose of corticosteroids, improving symptoms, and MRI and CSF findings. Toxicity associated with the short-course cyclophosphamide therapy (3–6 months) is minimal.[8] Cyclosporine at 5 mg/kg/day in two divided oral doses, in conjunction with prednisone, has been successful for refractory neurosarcoidosis.[132] Infliximab, a tumor necrosis factor (TNF-α) inhibitor has been successfully used in refractory neurosarcoidosis, including spinal cord dis-

Figure 6. Recommendations for treatment based on the authors' experience



ease.[2,3,133,134] When medical therapy fails, radiotherapy has been used with minimal side effects to treat spinal cord sarcoidosis.[135-138] Our practice to treat spinal cord sarcoidosis is outlined in Figure 6.

CONCLUSION

Spinal cord sarcoidosis is rare. Knowledge of its optimal approach to diagnosis and management is limited. We reviewed published reports and our experience with eight patients and derived algorithms for diagnosis and treatment of this rare disease. The diagnosis of the spinal sarcoidosis can be difficult. Treatment options are many, but they are not without side effects. A combination therapy is often helpful in controlling the symptoms and avoiding adverse effects.

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